Section for the Study of Disease in Children

President—Sir LANCELOT BARRINGTON-WARD, K.C.V.O., F.R.C.S.

[*May* 22, 1936]

Congenital Deformities of Mechanical Origin

By DENIS BROWNE, F.R.C.S.

THERE are certain disasters to which the body is liable—for instance, cuts, fractures, and burns—which could equally well be suffered by a lifeless model with the same physical qualities. There are others, such as bacterial infections and disturbances of growth, which can only affect living tissues. Now it is the purpose of this paper to argue that both these classes occur in the long list of congenital deformities.

The failure of purely vital processes is, of course, generally recognized in such conditions as hare-lip, achondroplasia, or branchial fistula. But I believe that as well as these there is a class in which the primary cause is something as mechanically simple and as independent of vital processes as a fracture of a bone. Just as in the fractured bone, there are, of course, secondary purely vital changes. But my argument is that the primary deformity of a club-foot could be reproduced in a fœtus made of putty, and that the underlying pathology of arthrogryposis would operate if the blood-vessels were formed of indiarubber instead of living cells.

The present teaching of the profession on this point is completely unsettled. The notion that congenital deformities may be caused by pressure *in utero* dates back at least to Hippocrates. But text-books on orthopædics merely give this suggestion a deprecating mention as a rather outmoded hypothesis; while certain workers of considerable authority on the subject, such as Middleton, flatly deny that anything of the sort can occur. To prove the question one way or another by direct observation is at present impossible, and it appears likely ever to remain so. In consequence I am reduced to a method that might possibly be used more in medicine than it is, the method of comparing what abstract argument shows to be the consequence of the granting of the hypothesis under test with what is actually found in real life. If the results of abstract inductive reasoning of this sort coincide with those of observation over a wide and complicated range, the truth of the hypothesis on which the reasoning was conducted is proved as nearly absolutely as most things can be in this world; and one can claim one more application of the razor of William of Ockham, one more general rule established, and in consequence one more diminution of the number of categories into which our observations must be placed. But it may be permissible to point out the main danger of working on these lines, a danger which I would call the "absolutist fallacy." This is the temptation to use the razor too enthusiastically; to argue that because a hypothesis is true over a certain observed range, it must therefore be true absolutely and universally. This way of thinking pervades all modern life so much that it is never named and seldom recognized. Darwin is a classical example of it, and I need not mention its political applications. But it is important to my case because so many people argue that if some deformities are obviously not caused mechanically, therefore none can be.

In stating my argument I first try to establish that the normal newborn baby shows obvious stigmata of mechanical compression. The importance of this is, of course, that if mechanical pressure is supposed to increase, these stigmata should increase in severity in proportion; and if it were to decrease, they should decrease too. Then I divide up the possible mechanical disasters that may befall the fœtus into arbitrary categories and discuss them separately.

(1) *Effects of Normal Mechanical Pressure upon a Fœtus in Normal Position*

(*a*) *The bending of the back.*—The fœtus begins as a straight plate but spends most of its intra-uterine existence huddled with bent back in a posture that obviously shows considerable pressure. In adult life there is a concavity in the lumbar region. In the uterus the pressure of the surroundings makes this same lumbar region the site of the maximum convexity into which the spine is forced [1].

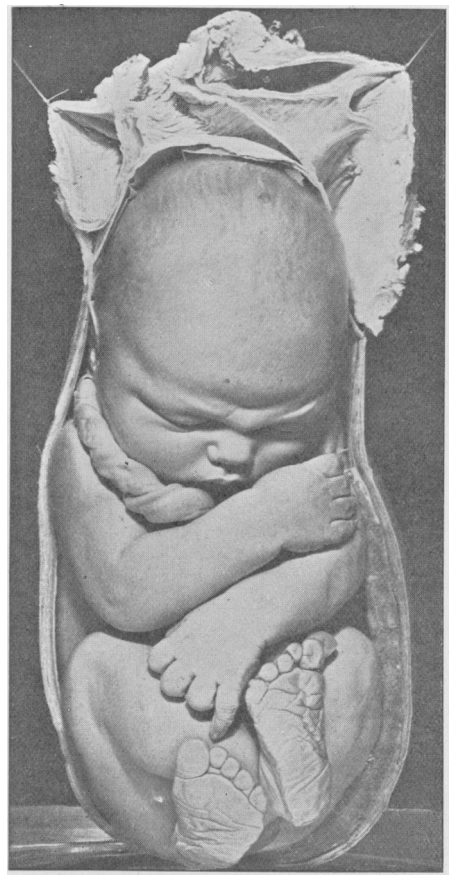

FIG. 1.—Fœtus in utero. This photograph of a preparation by Dr. H. R. Spencer is shown by kind permission of University College Hospital Medical School. Note how all the normal sites of pressure-dimples are over bony points in contact with the uterine walls. The way the feet take the pressure of the surroundings on their soles is most important.

(*b*) *The calcaneus position of the feet.*—It is not usually commented upon that the feet of a new-born baby are quite different in position and range of movement from those of an adult. They will go up into calcaneus with ease till the back of the toes touches the front of the shin. The reason is that *in utero* they take the pressure of the wall upon their soles. It is only necessary to study the illustrations in text-books of obstetrics to see that this is not generally recognized (fig. 1).

(*c*) *The dimples over bony points in contact with the uterine wall.*—These small pits are most important to my argument, as if they are caused in the way I maintain

their severity should give a good indication of the amount of intra-uterine mechanical pressure. I think they are due to the catching of the tissues between a sharp bony point and the uterine wall, and their consequently becoming compressed and adherent. Then when the pressure is released and the surrounding parts can stand up, they remain tied down and form small pits or dimples. Note that these never occur on otherwise suitable points that are not exposed to pressure, for instance, the anterior superior spine of the ilium, or the head of the clavicle, or the inside of the knee. They should also appear in abnormal situations according to abnormal positions of the fœtus.

(2) *The Effect of Normal Pressure and Development of the Fœtus plus Abnormal Position*

In this category come the only really curable conditions in the whole group of pressure deformities. Fortunately they also include the commonest ones.

(*a*) *Talipes equino-varus.*—An important part of the abstract argument is that the regions of the body most likely to be affected by mechanical pressure should also

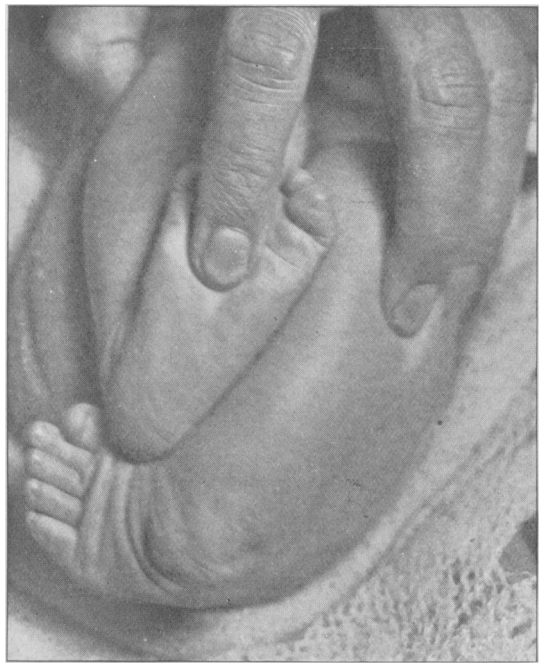

FIG. 2.—A left unilateral talipes held in the position assumed to have caused it. Note the thin and adherent skin over the area of greatest pressure in front of the external malleolus. The right foot is in normal position.

be found the most frequently deformed in real life. The parts most likely to get misplaced are the limbs ; and the arms are sheltered from pressure, by the overhanging head. Consequently it is the feet which one should expect to find far the most commonly deformed, and this by going in the easiest way they could go, by twisting inwards and so getting caught. The deformity which the orthopædic text-books state to be the commonest, talipes equino-varus, is the middle range (that is to say the most likely to occur) of an unbroken series of deformities all exactly corresponding to what one would expect from various degrees of this disaster. I have worked this out at length elsewhere [2] (figs. 2, 3, 4, 5).

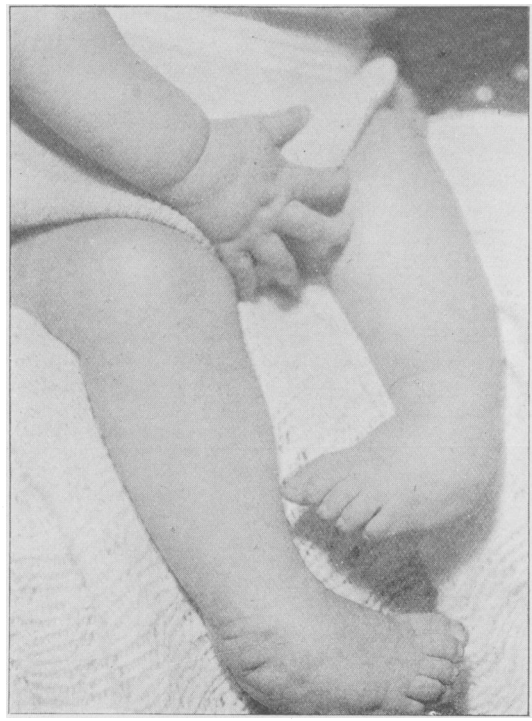

FIG. 3.—Double talipes, showing abnormal dimples over bony points on the outer side of the feet, in series with normal dimples over the knuckles. Note that the left foot is slightly more deformed than the right, having presumably been on the outer side in the cross-legged position.

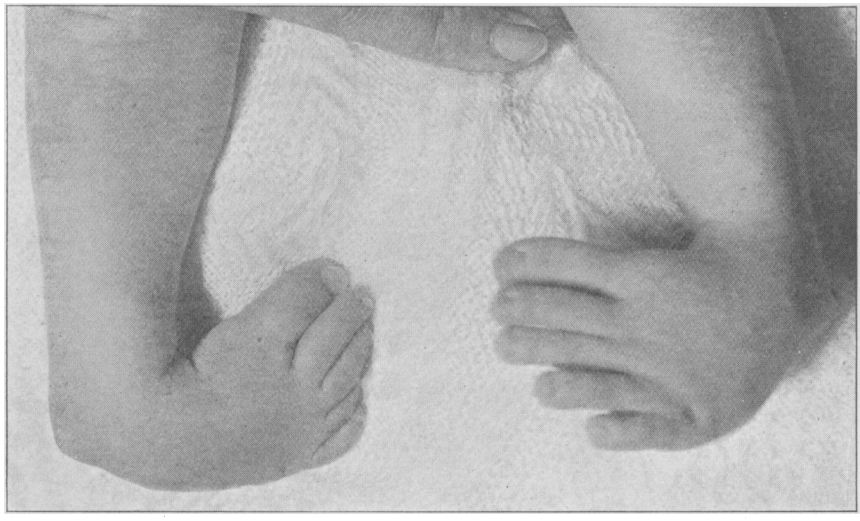

FIG. 4.—Extreme double talipes, showing abnormal dimples over the heads of the fifth metatarsals. In this case the right foot was the worse of the two. The muscles also, though not completely paralysed, were much below normal in activity. Although this is obviously merely a more severe degree of the same deformity shown in fig. 3, and usually called talipes equino-varus, the foot is in calcaneus rather than equinus.

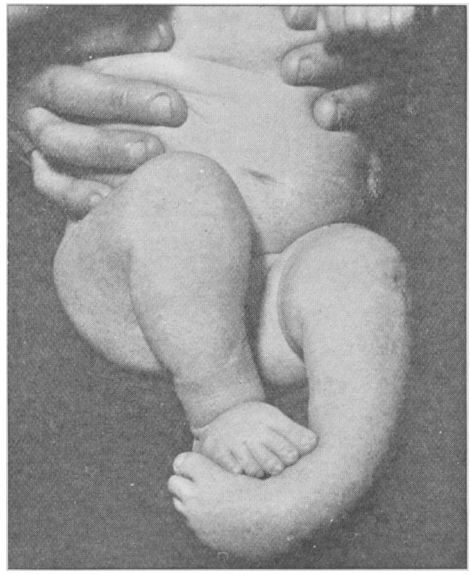

FIG. 5.—Left talipes, showing marked pressure-dimple over the external side of the head of the tibia. Note how well the curve of the deformed limb reproduces that of the uterus, and the way the limbs are naturally held in their original position.

(*b*) *Malposition of toes.*—Here is an example of this common deformity (fig. 6), showing also the first instance of another very important line of proofs for my hypothesis; the occurrence of mutual or corresponding deformities. The deformity

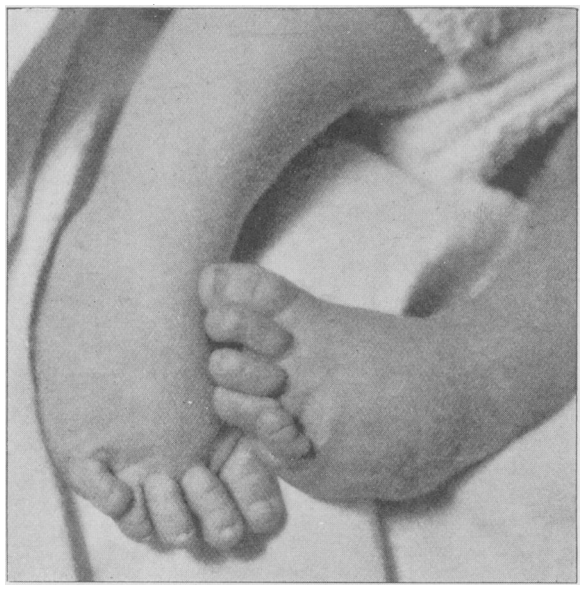

FIG. 6.—Double talipes with displaced little toes that fit into depressions on the back of the foot behind them.

of a disc of copper corresponds to the shape of the die that stamps it into a penny, and creates a strong presumption that the two have been pressed together. Similarly the fact that just behind these displaced toes there is a cavity that exactly fits them has similar implications of pressure.

(*c*) *Postural torticollis.*—This is a common deformity again, though usually self-correcting (fig. 7). The child is born holding its head on one side, and this position persists for several months, though there is no obvious failure of development beyond a flattening of the muscles of the compressed side. The photograph shows a most interesting case sent to me from Queen Charlotte's Hospital, with a note

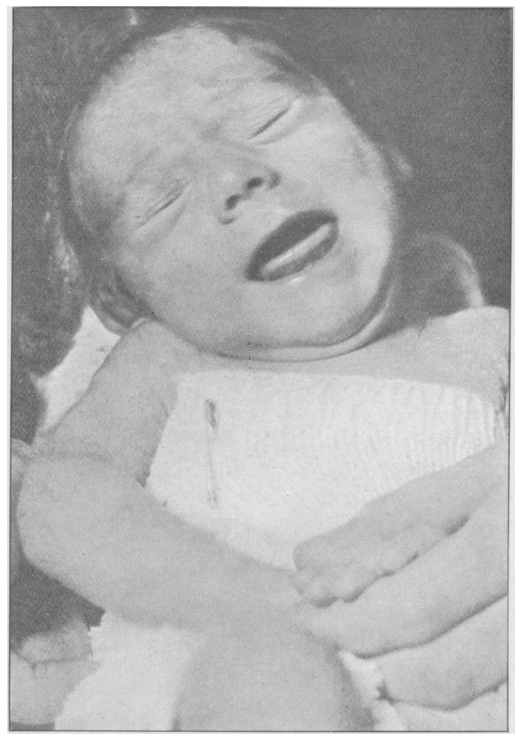

FIG. 7.—Postural torticollis. The head was persistently held in this position. The displaced lobe of the ear can be made out above the right shoulder, which is obviously not far from the emerging facial nerve.

that the obstetrician had actually felt the head to be jammed on one side while the child was *in utero.*

(*d*) *Displaced ears.*—Note in the same picture the displacement forward and upward of the ear, exactly corresponding to the way it is pushed against the shoulder. Quite a common condition.

(*e*) *Facial paralysis.*—In two of these cases I have seen transient facial paralysis, which it is tempting to explain as being due to pinching of the nerve between the shoulder and the mastoid process, on the analogy of crutch or operation paralysis. Of course this causation does not exclude the production of similar paralyses by forceps or by hæmorrhages.

(*f*) *Mutual deformity of feet and face.*—This is a single but most interesting case which, to my regret, I found before I had learnt the importance of cinematography as a record (figs. 8 and 9). Shortly after birth the child folded up with the greatest facility, thanks to flexed hips and hyper-extended knees, so that its compressed feet exactly fitted into a dish-shaped depression in the middle of its face. Compare with figs. 20, 21, and 22 (pp. 63, 64).

(*g*) *Acrocephalo-syndactyly.*—I put this condition here, though I suspect that there is usually an increase of mechanical pressure in it, because of its analogies with the preceding case. The associated deformities of the sides or front of the head and of the hands can be explained as mutual deformities on the assumption that the limb-buds grew too much upwards and landed the hands between the skull

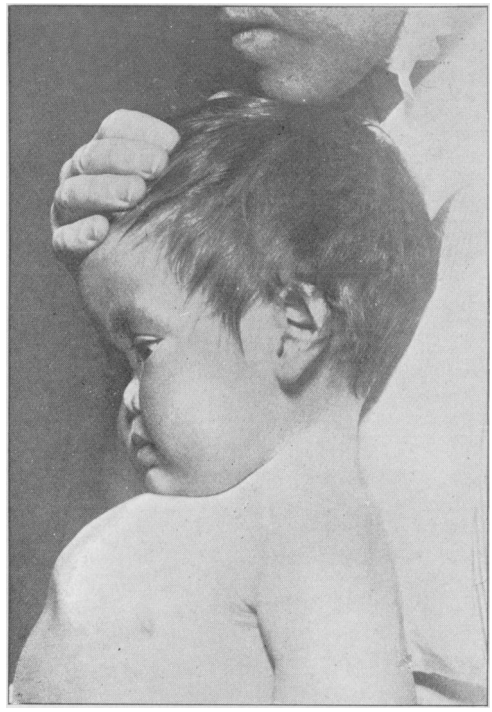

FIG. 8.—Face of a child who was folded up *in utero* with the feet fitting into the extraordinary dish-shaped depression in the middle of the face, across the bridge of the nose.

and the uterine wall. The reason why the development and separation of the fingers is imperfect is that this position can only be held by the growing up of the limb-buds, like the roots of a plant, before they have acquired the power of independent movement. They are not locked in position, as are the feet in talipes, and once the muscles have developed the hands would promptly be dragged back into normal position if they were to become displaced in this way. In some hundred papers that have been written on the subject this possible cause of the condition has not been mentioned, even to be disproved [3, 4, 5].

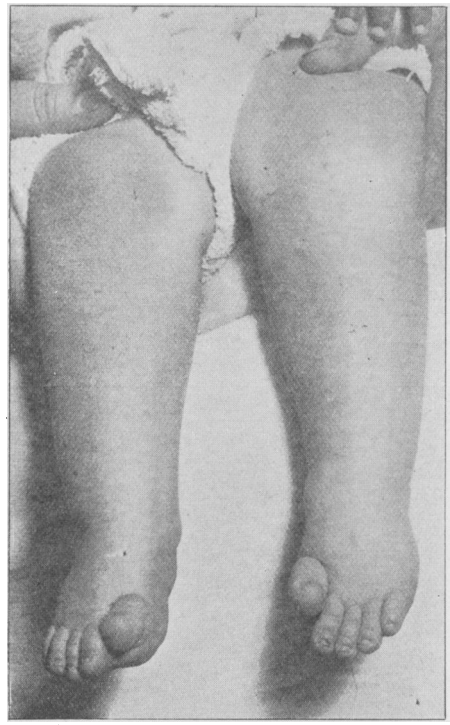

FIG. 9.—The feet of the same case as shown in fig. 8. Note the big toes driven back and folded up by contact with the bridge of the nose.

(3) *Normal Mechanical and Hydraulic Pressure plus Defective Development*

The defective development has, of course, nothing to do with mechanical influence of any kind.

(*a*) *Congenital dislocation of the hip.*—In the normal intra-uterine position there is obviously a thrust on the knee forcing the head of the femur downwards and backwards. Given an abnormally shallow acetabulum, why should this not dislocate the joint? If the joint is actually developed in a state of dislocation, as appears to be the most popular notion at the present time (apart from the difficulty of imagining how this could happen, given the method of formation of joints), it is curious that this disaster should happen only to the one joint that is exposed to pressure in the uterus, and that the displacement should invariably be in the only direction in which this pressure could force it. Since writing this I have found an ally in this theory of congenital dislocation of the hip in Professor Bauer of Vienna, who also, I am glad to say, appears to share my dislike of immobilizing these conditions [6].

I do not admit, by the way, that hyper-extension of the knee, or the displacement of the head of the astragalus in talipes, are true dislocations at all [7].

(*b*) *Congenital fracture of the tibia.*—Here there is almost always a faulty formation of the bones of the leg, usually an absence of the fibula. The invariable "scar" over the apex of the angle in the tibia is typical of a pressure-dimple. As to the angle itself, if it is not an effect of the bending of the presumably weaker leg over the other, it is curious that the invariable equinus position of the foot should correspond to the force necessary, and that the point of bending should correspond to the spot just above the heel, where the other shin would catch to form a fulcrum (figs. 10, 11).

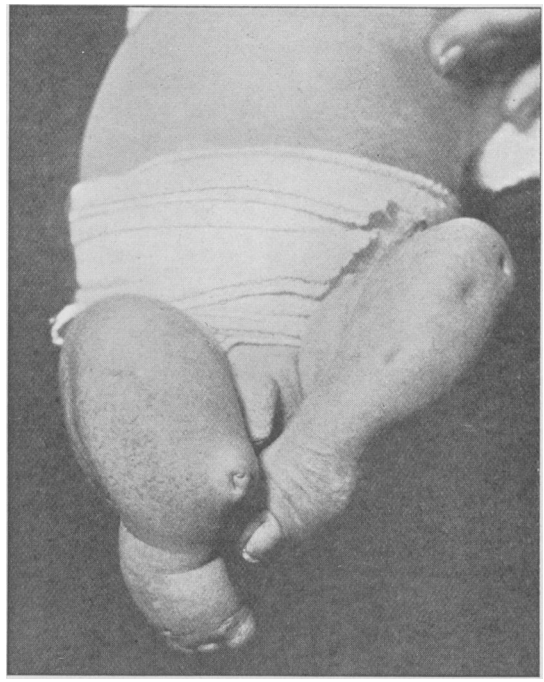

FIG. 10.—Case of congenital fracture of the right tibia, and deformity of the left leg. The fibula was absent on both sides. The dimple in the middle of the left shin, and a thin adherent patch of skin on the dorsum of the left foot, are both due to the pressure of the right leg when folded across it as shown in fig. 11.

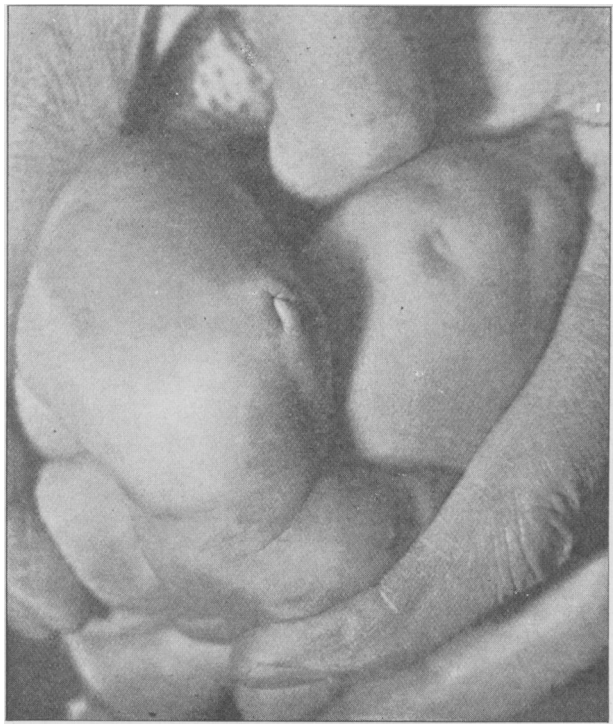

FIG. 11.—Same case as fig. 10, folded up in original position. Left leg shows a dimple over the edge of the external condyle of the femur and the outer side of the head of the tibia. There is a very good example of the classical one over the apex of the "fracture."

(*c*) *Club-hands* are usually deformities of this category, often with an absent ulna, and always with a pressure-dimple over the angle at the wrist.

(4) *Normal Hydraulic Pressure and Normal Position with Increased Mechanical Pressure*

Let us reason out what would happen in a hypothetical case of this. There should be an increase in the severity of those three stigmata of pressure which I have described in the normal infant.

(*a*) The spine should be not only bent, but should be compressed to such a degree that it collapses into secondary sideways bendings. It cannot bend very much farther than normal in the ordinary convexity backwards, because this is limited by the coming together of the head and limbs.

(*b*) The calcaneus position of the feet should be more marked, pushed to such a degree that there is actual deformity of shape, apart from difference of movement and posture from the adult condition.

FIG. 12.—Child who has been compressed in normal position. Note the very short back, with a lump over the lumbar region corresponding to a congenital scoliosis there. The normal dimples over the elbow and the outer side of the knee are exaggerated. Abnormal ones are to be seen over the first interphalangeal joints.

(*c*) The pressure-dimples should be in the same situations as normally, but should be more marked.

In addition a new set of troubles should be anticipated. If the limbs of an adult are kept closely confined for a considerable time, as for instance, after a fracture, degeneration of the tissues ensues, due to interference with the circulation. The muscles become atrophied and fibrosed, and the peri-articular tissues become infiltrated and stiffened, not infrequently permanently. (An excellent paper on these reactions by Mr. Watson Jones has just appeared in the *British Medical Journal* [8].) Finally, there might be nerve degenerations as well, due to pressure on exposed points

and analogous to crutch or post-operative palsy. It should be noted that the upper limbs, in their sheltered position, should be almost immune from mechanical pressure of this sort; so that it should be the rule to find the legs affected without the arms, but never the arms without the legs.

Now here in real life is the complete child we have evolved by abstract reasoning (figs. 12, 13). The spine is shortened by two acute scoliotic bends and the legs are exactly as postulated. The arms are normal. In contra-distinction to the common syndrome I am describing next, the sphincters are unaffected. The normal dimples are exaggerated in size and depth, and abnormal ones can be seen on the convexities of the phalangeal joints. Fig. 14 shows another example.

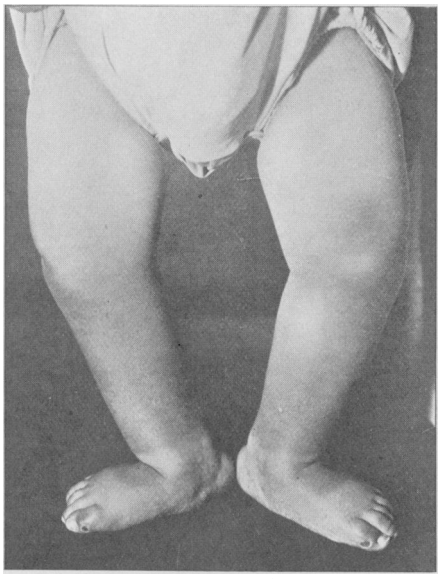

FIG. 13.—The feet of the same case as shown in fig. 12. Note the poor muscular development, the fixed calcaneus position, the knees still stiff in flexion, the abnormally long heels.

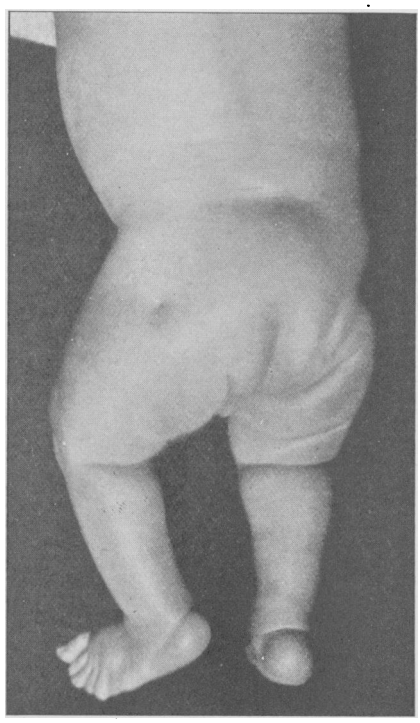

FIG. 14.—Another case of compression in the normal position. Note the very marked dimple in a not uncommon situation over the great trochanter of the femur, and abnormal projection of heels.

(5) *Effects of Normal Hydraulic Pressure plus Increased Mechanical Pressure plus Abnormal Position*

(*a*) *Club-feet of the usual type with muscle and joint degeneration.*—The methods of treatment which develop from considering the classic club-foot as a moulding deformity, and which I showed before the Section of Orthopædics in January, 1936, bring into sharp contrast those cases in which the muscles are normal (figs. 2, 3, 5, and 6, pp. 51-53), and those which with a similar deformity combine stiff joints and atrophied muscles (fig. 4, p. 52). The prognosis of the two types is totally different.

(*b*) *Atypical talipes with muscle and joint degeneration.*—This (fig. 15) explains itself.

(*c*) *A displaced finger showing arthrogryposis* confined to this finger alone, (fig. 16). According to my hypothesis the pathology of this finger is obvious. It is certainly

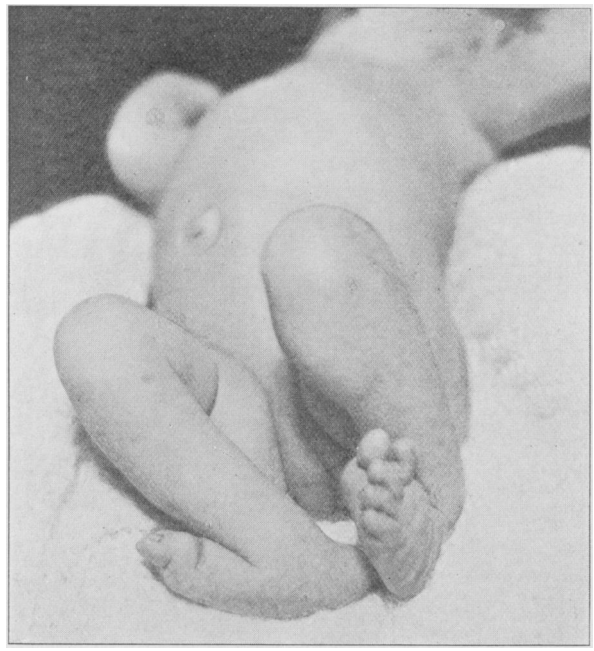

FIG. 15.—Mutual deformity of the feet in a case of severe mechanical compression. Note the extreme calcaneus of the right foot with exaggerated heel, and the adherent skin all over the dorsum of the left foot.

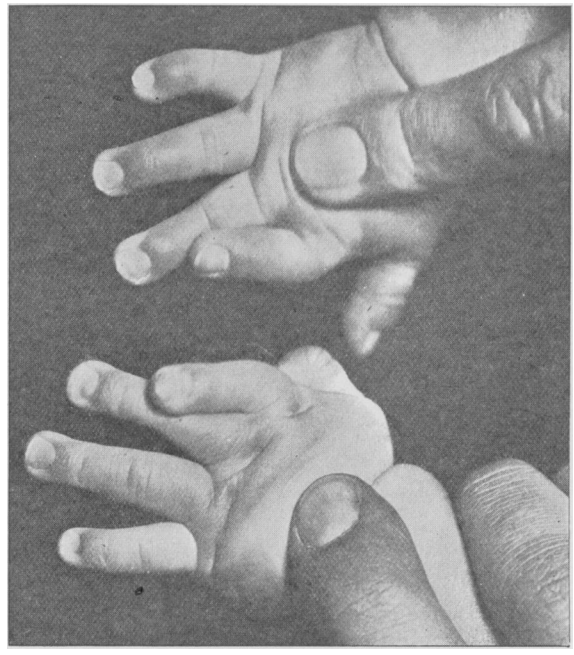

FIG. 16.—Double stiff forefinger, displaced over the dorsum of the middle finger. No active or passive bending of the interphalangeal joints was possible.

not easy to explain it as due to a primary muscular degeneration, as the wrist is perfectly free [9].

(*d*) *Spina bifida with talipes and degeneration of the muscles and joints of the legs.*—This most complicated and intractable deformity appears to me quite easily explicable under this heading. Let us imagine again what would happen if right at the start of fœtal life, even earlier than it was assumed in the previous instance, the relation between the expanding fœtus and its surroundings was disturbed, so that the bending of the back was exaggerated from its very beginning. At this time when the fœtus is barely 2 mm. long, the edges of the neural groove are joining to form the neural tube. Any extra bending would make the sides of this groove gape at the point of maximum curve, that is, the lumbar region. Accordingly faulty fusion would be likely, with the formation of a spina bifida at this point [1].

A second result of this extra bending would be the tilting upwards of the buds of the lower limbs, so that instead of the developing legs folding normally in front of the belly, they would stick straight up towards the head, and the feet would be forced

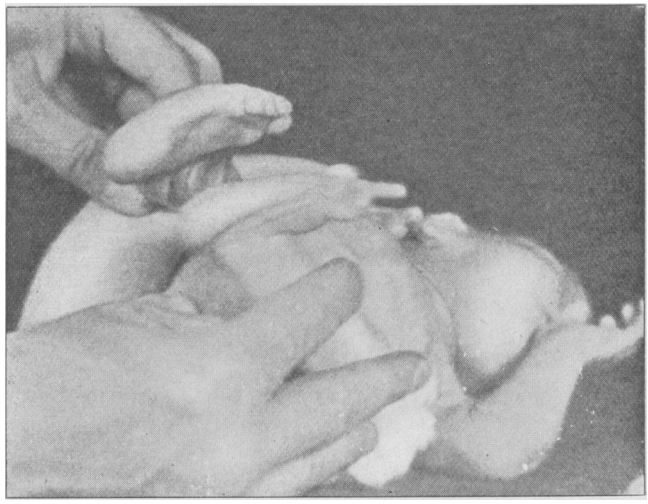

FIG. 17.—Mutual deformity of feet in a case of talipes with spina bifida. (From a cinematograph film, which shows vividly the way the feet correspond.) The knees are not in the more usual hyper-extended position.

into the uterine wall. The result of this would be flexed hips, hyper-extended knees, and irregular talipes; the only thing constant about the shape of the feet being that on one side or the other there should be a convexity corresponding to the uterine concavity.

Muscle degeneration and joint stiffening would be the same as in the previous case described. But there would be a new paralytic element in the damage to the nerves at the point of faulty development of the spinal cord. The result of this would be paralysed sphincters of the bladder and rectum, with the relaxed sphincter ani in a typical position on the surface of the buttocks, the normal cleft at the bottom of which it lies not developing because of the paralysis of the levator ani.

Of course the extended position of the legs is not invariable. It is not uncommon to find that they have been compressed in either the normal or the club-foot positions. But it is the commonest.

Here again are two actual examples of our abstraction (figs. 17 and 18) exact in every detail, and to be duplicated only too easily in any children's outpatient

department. Note the exceptionally good example of mutual moulding of the feet in fig. 17. Fig. 19 is a similar case.

(*e*) *A similar case to* (*d*), with mutual deformities of the head and feet (figs. 20, 21, and 22). The pictures are self-explanatory, and the analogy with acrocephalo-syndactyly and figs. 13 and 14 (p. 59) is obvious. There was a large lumbar spina bifida which does not show in the photograph.

(*f*) *A dimple over a displaced coccyx* is a minor confirmation of this hypothesis which can be not infrequently found (fig. 23, p. 65).

(*g*) *Talipes with dislocated hip and defective development* is a syndrome that few orthopædic surgeons can have been fortunate enough to escape meeting. It is hardly necessary to point out how it fits into this category.

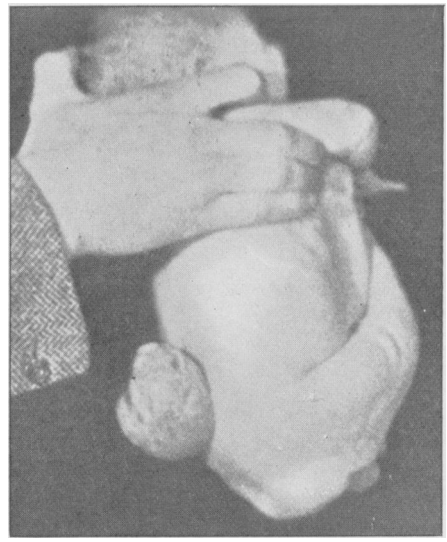

FIG. 18.—Case of spina bifida and talipes with hyper-extended knees. (From a cinematograph film.) Note the way one leg crosses the other. They can only be made to fit snugly in this way.

(6) *Effects of Pressure of Amniotic Bands*

I have seen only one case which I feel inclined to attribute to this. In it the effect was exactly as if on a face modelled in soft clay a handkerchief had been pulled across nose and cheek bones, interfering with ossification and dragging down the lower eyelids into a gross deformity. There was an interesting analogy in this case to the formation of lumbar spina bifida which I have mentioned, in that the head was flattened from front to back and a cleft palate was present, perhaps due to the increase in distance which this flattening caused between the joining palatal processes. There is also a reported extra liability to cleft palate in cases of acrocephalo-syndactyly which might possibly have a similar origin.

(7) *Effects of Spasmodic Muscular Pressure*

Fig. 24 (p. 65) shows a full-term baby fixed in the position caused by spasmodic contraction of the uterus. It gives a good idea of the force of such a contraction, and shows the way in which the normal position of the fœtus can be disarranged in the stress of birth, especially when the liquor amnii has escaped. It is surely not difficult to imagine that certain troubles can be produced in this way.

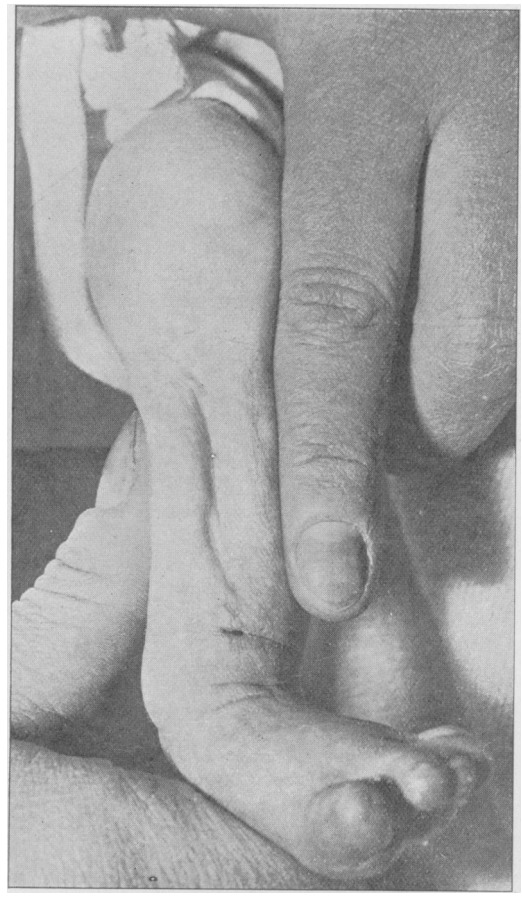

FIG. 19.—Compression dimple over the shin from a similar case to that shown in fig. 18. It was caused by the pressure of the other leg over it in a crossed position. There was no corresponding mark on the other leg.

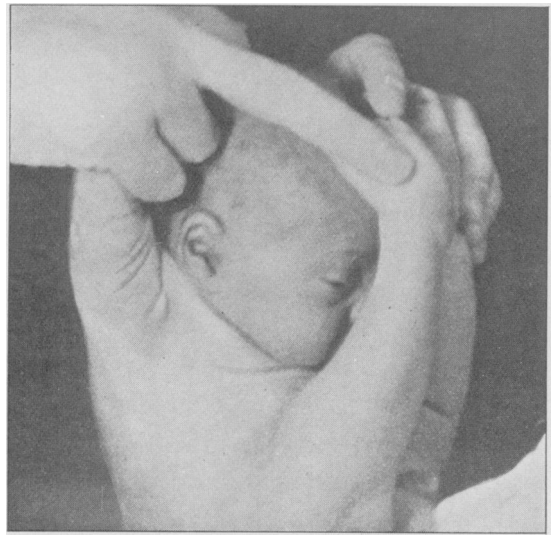

FIG. 20.—Case of large lumbar spina bifida and mutual deformities of feet and skull. This picture (from a cinematograph film) shows how the flexed hips and hyper-extended stiff knees allow the feet to come naturally against the forehead. The arms can quite easily be folded under the chin behind the legs while they are in this position.

(*a*) *The formation of a sternomastoid tumour*, with subsequent torticollis from shortening of that muscle, is, according to Middleton, a sequel of venous stasis caused during compression at birth. On the analogy of Volkmann's ischæmia this seems to me extremely likely.

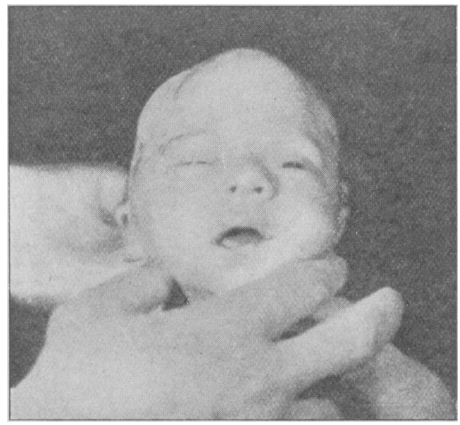

FIG. 21.—Head of same child as shown in fig. 20. Showing depressions on either side of forehead exactly corresponding to soles of feet.

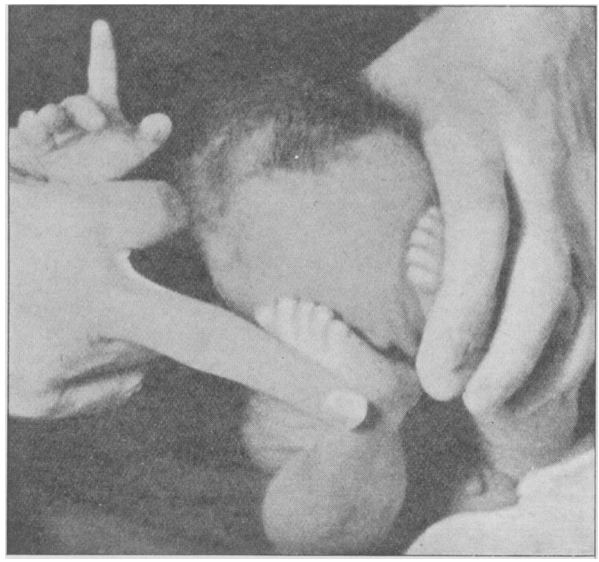

FIG. 22.—Top view of same child as shown in figs. 20 and 21, showing how feet fit depressions in forehead. Compare with figs. 8 and 9 (pp. 55, 56).

(*b*) *Dislocation of a normal hip.*—I have seen a baby which had a very easy and uncomplicated birth conducted under skilled observation. A week or so later the child was found to have a recent dislocation of a normal hip, which on being reduced gave a perfect result. There is, of course, no proof of what actually happened, but

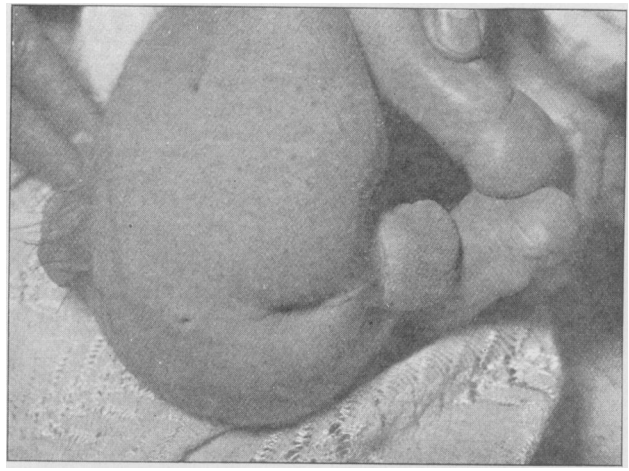

FIG. 23.—Dimple over sharp tip of a displaced coccyx in a case of talipes with spina bifida. Compare with "scar" over apex of bend in tibia in fig. 11 (p. 57).

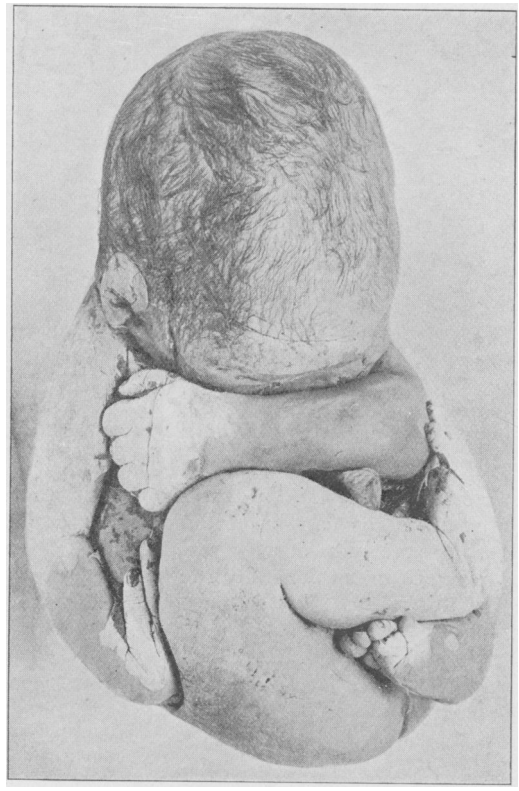

FIG. 24.—Extreme compression of full-term fœtus during birth. Note disarrangement of position of right hand and left foot, without deformity.

it seems to me most likely that the hip was driven out by a muscular spasm pressing the knee straight backwards. This straight drive of force along the axis of the bone would not tend to break the femur, but violence of the wrenching or dropping type would surely have produced the common birth fracture.

(*c*) *Fracture of tibia with backward angling.*—I have had another case in which, following persistent and violent attempts at abortion, a baby was born normal except for an acute backward bend in the tibia (fig. 25). The tibia folded up as the picture showed, and had the expected dimple over the convexity of the bend. After an osteotomy it behaved exactly like a malunion of an ordinary fracture, and it promises to give an excellent result. If this deformity was not mechanically produced by a contraction of the uterus, I can think of no other explanation. It certainly was not due to muscular degeneration, as there was none. Note the analogy with congenital angulation of the tibia.

(*d*) *Pressure sores on sole of foot.*—Another strange case which I put in as a curiosity, without insisting too much upon the explanation, is one which was seen by

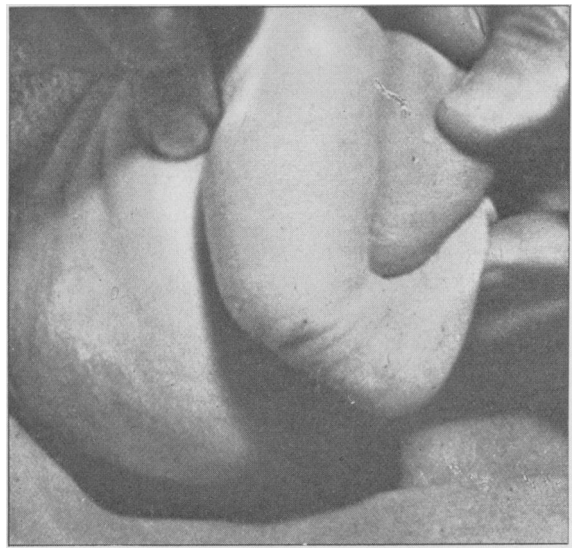

FIG. 25.—Fracture or bend of tibia and fibula in newborn baby. Note dimple over apex of bend, and how the position of this exactly corresponds (though pointing the opposite way) to that in fig. 10 (p. 57).

Dr. Bernard Schlesinger, who very kindly gave me this photograph (fig. 26). We all know how quickly pressure sores can be produced upon a baby's skin, and as this child was born with what exactly resemble pressure sores in exactly the place where uterine pressure according to me is borne by the feet, I thought this might be worth mentioning. In their subsequent history they behaved exactly in accord with this suggestion of their nature.

(*e*) *Birth fractures.*—I do not know enough about obstetrics to be sure whether some birth fractures are produced by spasms of the uterus rather than by the strong arm of the accoucheur.

(8) *Effects of Increased Hydraulic Pressure*

The hydraulics of the fœtus are of an extreme complication. But the important point to my argument is that the pressure of the amniotic fluid, though all pervading, is yet elastic; it is not a rigid fixed quantity, as it would be at the bottom of a tank,

but varies with the tension of the amnion and the uterus. This pressure could be increased in two ways, first by the amount of fluid being too large, and secondly by the container being too small. Probably the former is the commoner; it is difficult to imagine sudden hydramnios without an increase in hydraulic tension, as in an acute hydrocœle. But I believe the latter also occurs, and in one of the cases in which I suspected it there was the very suggestive history that throughout the later stages of the pregnancy the mother was in constant pain, quite unlike anything she had felt with a previous child.

With such an increase one would expect that the waves of blood sent into the fœtal circulation from its own heart and the maternal one would be very rapidly diminished by the all-pervading elastic pressure. Consequently only the muscles and viscera close to the impulse would get a full supply, and the limbs would be in a condition of reduced arterial supply combined with venous stasis. The degeneration of muscle and stiffening of joints already described would occur just as before, but the nerves would be much less liable to harm than they are in mechanical pressure.

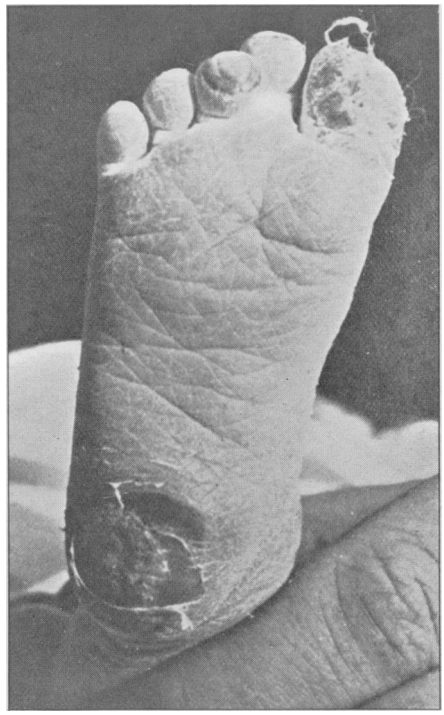

FIG. 26.—Foot of new-born baby, showing marks suggesting pressure sores.

Consequently the muscles which are left in contraction by whatever position the fœtus is in should retain some power, and might even lead to typical deformities by their contraction. Where there is hydramnios two further results should follow. First, the extra space should give the fœtus room to assume, and to stiffen in, positions that would be impossible in the normal cramped surroundings; for instance, the hips might be extended. Secondly, the less close contact of the uterine walls in the hydramniotic cases should cause less marking of pressure dimples than normal (fig. 29, p. 69).

Fig. 27 shows an example. The child was born rigidly stiff in the position shown, with extended hips. The deformities of the feet and hands are what would be expected from a combination of the position of the limbs, the impact of the uterine walls, and muscular contraction. There was a history of hydramnios. Fig. 28 shows another baby with the same characteristics, except that the thighs originally were flexed and the knees extended, as in Sheldon's case [10]. Note the deformity of the hands; it is obviously much more due to muscular contraction than to moulding.

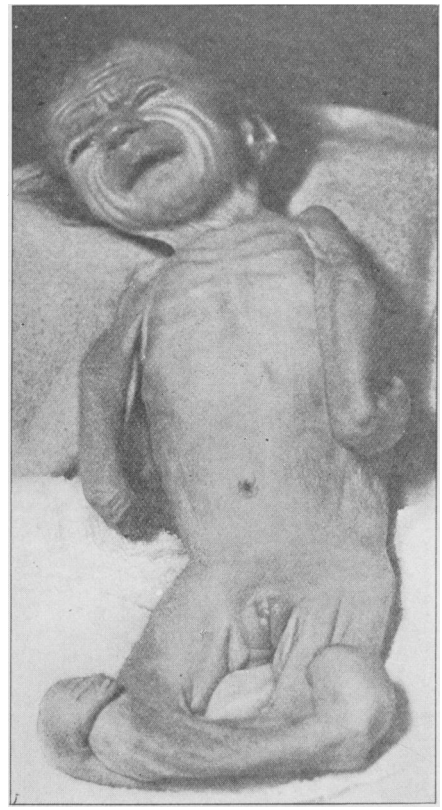

FIG. 27.—Case of "arthrogryposis." Note extended hips.

Finally the absence of even the normal dimples of the outer side of the elbow, which should be expected in such cases, is well shown in fig. 29.

A most suggestive parallel can be drawn between these cases and a disease of sheep [11]. In certain flocks lambs are born with stiff limbs set out at abnormal angles, the state of the muscles and joints being much as described. But the interesting thing is that the shepherds can foretell the appearance of one of these lambs by the undue size of the mother's belly. This increase in size must be due to hydramnios; this hydramnios must cause increased hydraulic pressure. Increased hydraulic pressure is what I postulated for the production of these cases of "arthrogryposis."

FIG. 29.—Case of "arthrogryposis." Note complete absence of normal moulding at elbow.

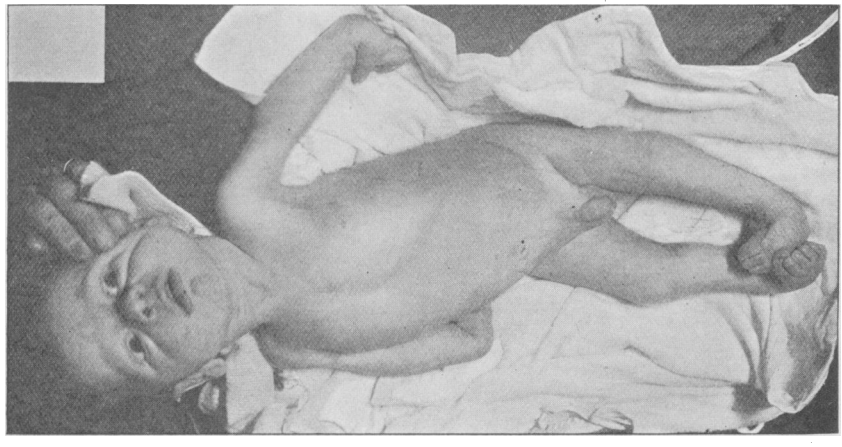

FIG. 28.—Case of "arthrogryposis." Note absence of dimples.

(9) *Deformities Not Due to Mechanical Causes*

This section is of great importance, not so much to my argument as to the acceptance of it. This is because of the use of the "absolutist fallacy" as a means of attack, the objection that if it does not explain condition A it cannot explain condition B.

(*a*) *Local failures of formation*, the missing fibula in congenital fractures, &c.

(*b*) *Ring constrictions of the extremities*, for reasons given by Streeter [12].

(*c*) *Sprengel's shoulder.*—I regard this as a failure of migration, analogous to undescended testicle. The shoulder is formed high up and should descend. If it does not the muscles above it cannot develop and one would expect to find them atrophied, as in fact one does. There are no pressure-dimples in a place excellently fitted to show them; and it is difficult to imagine a one-sided thrust of this sort being delivered by any available mechanism.

(*d*) *Achondroplasia and various forms of dwarfism.*—These are attributed by Jansen [15] to selective action on certain groups of cells by temporary increases of hydraulic pressure during fœtal life. There is no proof that these cells are sensitive in this way, nor that these increases occur. In addition, it would surely be expected that it would be common to find children showing all these different failures of development on top of one another in cases in which there had been an increase of pressure throughout pregnancy.

(10) *Alternative Explanations of Some of the Conditions Described*

(*a*) *Degeneration of muscle.*—The suggestion that this condition is a primary spontaneous one has two obvious weaknesses. First, it assumes a totally new pathological condition. Second, it does not explain the associated conditions; the stiffening of the joints, the deformities of the feet, and the presence of pressure-dimples when the legs alone are affected, and their absence in many cases when the arms suffer too.

(*b*) *Dimples* are explained by Ombredanne [13] as being due to intra-uterine ulceration. There is this to be said for it, that cases are on record where there have been granulating areas over such points. However, I would simply class these as pressure-sores.

Middleton says that they mark the spots where bursæ will develop in later life, but gives no reason why they should. I do not agree that the situation of a knee dimple coincides with that of a housemaid's knee.

Lovett and Jones say, without explanations, that they are due to amniotic adhesions.

Talipes.—(*a*) Arrest at certain stages of development is an explanation of talipes equino-varus that is very popular in Germany. An argument against it is that positions with any real resemblance to talipes do not occur in normal fœtal life, though of course it is very possible that talipes feet have never at any time been in correct position. Apart from this it gives no explanation of atypical talipes.

(*b*) Nervous action is often suggested, especially in cases combined with spina bifida. Now the nerves could only act through the muscles and muscles cannot produce accurately a simple club-foot, let alone any of the others.

(*c*) Brockman suggests that a primary dislocation of the head of the astragalus is the cause of talipes equino-varus.

Congenital fracture of the tibia.—Middleton suggests that this is due to a primary degeneration and shortening of the calf muscles. He does not say why the tibia bends sharply at one point instead of along its length like a bow. It seems to me quite impossible to explain my case of backward bowing by this. Admittedly the muscles of the calf are atrophied and contracted, but seeing they have been under pressure and that the heel in these cases is about opposite the middle of the tibia,

I think this is only to be expected. Compare the muscles above a Sprengel's shoulder.

Arthrogryposis.—This is explained as being due to a primary muscular degeneration. I have seen several cases in which it is confined to the fingers alone, with and without displacement. This is easily explainable if it is assumed to be due to pressure, but a primary muscular atrophy would give a very different picture.

(11) *Arguments against the Mechanical Causation of Deformities*

(*a*) Little [14] says that pressure cannot be the cause of talipes because it has been found in fœtuses of three and four months old.

(*b*) It is argued that spina bifida cannot be due to excessive bending of the fœtus because this must occur at such an early stage. I cannot understand this objection, but it is so constantly made that I mention it.

(*c*) Finally there is what is not so much an argument as a statement; the assertion that the folding up of mechanically compressed babies in abnormal ways is the result of the deformity and not its cause [9]. As far as I can grasp it this means that the feet in fig. 17 (p. 61) have not been shaped by being pressed together, but that they fit together because they have spontaneously assumed that shape.

For most of the photographs on which my argument depends I am indebted to Mr. Derek Martin, of the Pathological Department of the Hospital for Sick Children, Great Ormond Street, London. Those who have tried themselves to photograph special points on small infants will understand what skill and patience has been needed.

REFERENCES

1 "Gray's anatomy," 26th Edition, p. 60, fig. 79.
2 BROWNE, DENIS. *Lancet*, 1934 (ii), 969.
3 HILL, N. *Proc. Roy. Soc. Med.*, 1929, **22**, 72.
4 SMITH, J. *Ibid.*, 1930, **24**, 2.
5 PARK, E. A., and POWER, G. F. *Am. Journ. Dis. Child.*, 1920, **20**, 235.
6 BAUER, F. *Lancet*, 1936 (i), 1057.
7 BROCKMAN, E. P. "Congenital club-foot," London, 1930.
8 JONES, WATSON. *Brit. Med. Journ.*, 1936 (i), 925.
9 MIDDLETON, D. S. *Brit. Journ. Surg.*, 1935, **22**, No. 88, 696; *ibid.*, 1930, No. 70, 188; *Edin. Med. Journ.*, 1934, **41**, 401.
10 SHELDON, W. *Archives Dis. Child.*, 1932, **7**, 117.
11 ROBERTS, J. A. F. *Journ. Min. Agric.*, 1926, **33**, 795; *Journ. Genetics*, 1929, **21**, 57.
12 STREETER, G. L. *Contr. Embryolog. Carn. Instit.*, Washington, 1930, **22**, 1.
13 OMBREDANNE, "Chirurgie infantile," Paris, 1932.
14 LITTLE, W. J. "On club-feet," London, 1839.
15 JANSEN, M. "Faiblesse de croissance et nanisme congénital," Paris, 1926.

CASES

Persistent Acidosis in an Infant: Cause not yet ascertained.—REGINALD LIGHTWOOD, M.D., N. F. MACLAGAN, M.D., and J. G. WILLIAMS, M.R.C.S., L.R.C.P.

P. H., female, aged 14 months; breast-fed until nine months old; did well until 7½ months, when she ceased to gain weight. At the time of weaning on to a suitable mixed diet of milk, groats, broth, &c., she began to vomit at irregular intervals in a somewhat projectile manner. In four weeks she lost 2 lb., and was brought to the Hospital for Sick Children and seen by one of us (R. L.). The mother reported constipation, poor appetite, and slight spasmodic movements of the right arm and leg.

Examination (1.2.36).—Aged 10 months, weight 15 lb. 10 oz. Apathetic, good colour, fair nutrition, suspicion of dehydration, muscles surprisingly hypotonic, trace of albumin in urine.